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CASE REPORT

Tongue pearl: A novel technique for treatment of an infant with median facial cleft and microcephaly ☆

Sherif Essam Zahra [a,*], **Mamdouh Aboul Hassan** [b]

[a] *Department of Orthodontics, Faculty of Dentistry, Suez Canal University, Ismailia, P.O. Box 3414, New Campus, Ring Road, Egypt*
[b] *Abu El Reesh Specialized Hospital for Children, Cairo University, Egypt*

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KEYWORDS

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Abstract This case report illustrates the presurgical treatment of a patient having a median facial cleft and microcephaly, using a guidance appliance. The appliance was custom designed and modified with a pearl-like acrylic structure attached to its lingual surface for pushing the extremely protruded tongue back to its normal position to facilitate anesthesia and surgical lip closure. Total treatment time was 5 weeks. Regaining normal tongue position, in turn, facilitated both intubation and extubation, preventing the postoperative respiratory distress the authors had experienced with similar cases.

1. Introduction

Midline facial cleft is an extremely rare entity, occurring in probably less than 1 per 100,000 births (Kawamoto and Patel, 1998). It may present with a wide spectrum of features, including hypertelorbitism, V-shaped frontal hairline, bifid cranium, broad nasal root, bifid nose, median cleft of the upper lip, and median cleft of the premaxilla (Urata and Kawamoto, 2003). The midline, or median, facial cleft is most commonly known as a 0–14 cleft, as classified by Tessier (1976). Based on the author's experience with similar cases (median facial cleft with severe tongue protrusion coupled with microcephaly), there are complications typically associated with the emergence and maintenance of a stable upper airway during intubation, ventilation, and extubation. In addition, there is frequently upper airway respiratory distress immediately postoperatively, wherein the child must be intubated for 48 h. This is explained on the basis of faulty tongue position, postoperative edema of the tissues following lip closure, and stresses of the attached muscles pulling the tongue backward, all leading to dyspnea.

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* Corresponding author. Address: 23 Kirdahy Street, Rushdy, Alexandria, Flat 5, Egypt. Tel.: +20 12 3115154.
E-mail addresses: sherife@odont.uio.no (S.E. Zahra), aboulhassanm2@gmail.com (M.A. Hassan).

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In a study conducted to measure the clinical impact of microcephaly among children with developmental disorders, it was found that respiratory distress and intraventricular hemorrhage showed the strongest correlation (Watemberg et al., 2002). The aim of this case report is to describe a new appliance and treatment approach to push back and normalize the position of an extremely protruded tongue in an infant with median facial cleft and microcephaly.

2. Case report

2.1. Patient history, clinical examination, and diagnosis

The patient was referred to the pediatric plastic surgery department of Cairo University's specialized children's hospital at the age of 11 months with a median cleft lip. She was the first daughter of a healthy mother and father who were in their mid-20s. There were no complications during delivery, as stated by the parents. The mother and father reported no medical problems or history of facial clefting in the family, and there was no reported consanguinity. Medical reports of the child revealed that she was born with microcephaly due to a brain cyst. Initial examination (Fig. 1A and B) revealed a bifid nose with mild asymmetry of the nasal apertures and wide median cleft lip (CL) and alveolar process. Hypertelorbitism was noted. No other anomalies were found outside the head and skull. The patient's tongue protruded outward and upward, occupying the cleft most of the time. Based on these characteristic observations, the median facial cleft was classified as Tessier 0. Surgical closure of the lip was intended to improve the patient's appearance, to be followed by nasal repair at a later age.

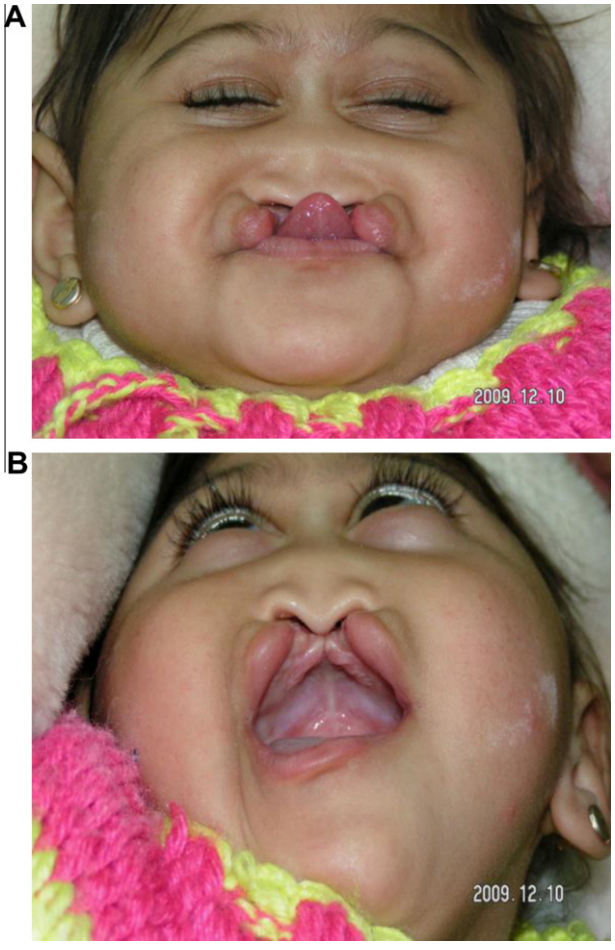

Figure 1 (A) Pre-treatment facial photograph showing wide median cleft of the upper lip and severely protruded tongue. (B) Pretreatment intra-oral view showing cleft alveolus.

2.2. Appliance and technique

To facilitate lip surgery, a maxillary appliance was fabricated to push the tongue back to its normal position; a passive acrylic plate (Fig. 2) with stainless steel wire outriggers that relied on the facial musculature to seat the plate. This was a primary concern as the child was almost 1 year old and could remove the plate. In addition, plaster tapes across the cheek were used for further stability of the appliance. A modification to the appliance in the form of an acrylic pearl structure on the anterior part of the plate was constructed to individually suit the case needs. The seated plate together with the pearl prevents the tongue from thrusting between the alveolar segments and exerting any widening influence. Also it helps to normalize tongue-tip placement and activity by forcing the tongue to return to its normal position. For the pearl to function correctly, its size and position are of prime importance. It should not be too big or too far posteriorly, otherwise it will not allow the tongue to regain its normal position. The parents were instructed to keep the plate in position 24 h a day and remove it only for cleaning after feeding (Fig. 3). They were also told to tape across the gap between the upper lip segments after the child adapts to the appliance. They reported that during the first week, there was some difficulty with wearing the appliance and rejection but afterward, the child got used to it.

2.3. Treatment progress

A dramatic improvement of the tongue position was noticed during the third week of appliance wear (Fig. 4); however, it

Figure 2 Modified acrylic plate including stainless steel wire outriggers.

Figure 3 Patient wearing the modified appliance.

Figure 4 Post-treatment facial photograph showing improvement in tongue position.

Figure 5 A facial photograph of the patient after lip closure.

was decided that the child should continue wearing the appliance for another 2 weeks to retain the new tongue position. Surgical lip closure was scheduled (Fig. 5). The patient showed both a smooth intubation and recovery from anesthesia, and she was followed up for any signs for respiratory distress. She was dismissed 2 days postsurgery and the appliance wear was discontinued.

3. Discussion

Unfortunately, evidence-based care for cleft patients is lacking, and treatment decisions are based on the empirical experience of the individual professional or on anecdotal reports of success of a specific treatment modality.

Birth defects of the face are often some of the most disturbing because we usually identify and differentiate people by facial appearance. Because median facial defects can include a median CL, it is important to distinguish between a median CL and an ordinary CL (both of which may also include a cleft palate). An ordinary CL is one of the most common congenital malformations, having an incidence of about 1 in 700 births (combining isolated defects with those associated with other anomalies). They are linear defects that extend from 1 of the nostrils down through the lip but are lateral to the philtrum. These clefts can be bilateral, extending down from each nostril; however, the tissue of the philtrum in the midline is still intact. A median CL involves the middle of the upper lip (or the area of the philtrum). This defect has a different pathophysiology. It is caused by lack of the developing maxillary bony processes to extend medially and join at the midline below the nose. This median CL anomaly (if not an isolated defect) has been associated with only 2 disorders: holoprosencephaly (a severe anomaly of the brain) and median facial plane defects (or frontonasal dysplasia) (De Meyer, 1967).

The increased incidence of anesthesiological complications in children with clefts can be attributed to various factors, among them developmental anomalies and anatomical features such as micrognathia, macroglossia, and jawbone hypoplasia. Furthermore, in cleft lip and palate patients, the anomaly requiring surgery can be associated with one of 150 different syndromes or nonsyndromic abnormalities (Ward, 1987; Hujoel et al., 1992).

Individual appliances may be necessary for the treatment of Tessier 0 median facial clefts due to a protruded tongue that can complicate anesthesia, hinder timely lip closure, and cause postoperative respiratory distress. In our protocol of treatment, we managed to achieve good results with a modified presurgical appliance. The challenge was to normalize tongue position to facilitate intubation and lip closure and to prevent the postoperative upper airway distress that had occurred with previous cases. An important goal is to maintain well-defined protocols for the management of the different malformations. This is critical in light of the pressing need for proper clinical trials involving large samples of patients. There is a growing body of evidence that collaboration should take place between craniofacial teams with a history of high volume and with an excellent track record for care of patients with craniofacial malformations. This approach will ensure a successful delivery of evidence-based care in future.

Ethical Statement

The work described in our article have been carried out in accordance with the Code of Ethics of the World Medical Association. The technique described in this article has been approved by the Ethical Committee in Cairo University Specialized Children Hospital. Also, this work was done after getting the approval from the parents of the child and agreed on publishing the child's photographs.

Conflict of interest

No conflict of interest declared.

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